CONGENITAL HEART DISEASE IN YAKIMA 2017

How to manage infants and children with congenital heart disease in central WA surrounded by mountain passes in winter and forest fires in summer

CENTRAL WA PEDIATRIC CARDIOLOGY SERVICES

- CLINICS 3/WEEK CHILDRENS VILLAGE YAKIMA 3/WEEK TRICITIES 1-2/WEEK WENATCHEE
- 40 pats/week 2000 per year emergency add-on open spot every clinic

- ECHO 800-1000/yr HOLTER/EVENT MONITOR FETAL ECHO 10/month

- REFER SCH FOR diagnostic/ interventional/EP CATH 65/yr and OHS 40-50 /yr

- OTHER SERV in (chi vill) feeding/neurodev/fam services/orthoped/GI/GENETIC
- (tricities) nephro/uro/gen surgery telemed derm
- (Wenatchee) ADCHD endocrine

SEATTLE CHILDRENS SERVICES

- OPEN HEART SURGERY 400-450 /yr 4 surgeons (mary bridge hospital Tacoma)
- Diagnostic/INTERVENTIONAL CARDIAC CATH 4 providers
  PV/ ao valve/coarct balloon PDA/ASD device coA/ductal stent
- ELECTROPHYS/ABLATION
- TRANSPLANT/HEART FAILURE 3 providers
- PAH/CPET
- MRI CT 3D reconstruction for planning of OHS
- Genetics/22 q 11 clinic / MD clinic / vascular anomaly clinic
- ADCHD 3 providers

PEDIATRIC CARDIOLOGY AND CARDIAC DISEASE IN CHILDREN AND ADOLESCENTS

L-R SHUNTS: VSD ASD PDA AVSD (AVC /ECD)

R-L SHUNTS : TOF D-TGA Tri ATRESIA PA-VSD PA-IVS TRUNCUS TAPVC EBSTEINS

OBSTRUCTIVE LESIONS: PS CoA HPLHS AS (subAS supraAS)

POSTINFECTIOUS : ARF/RHD KAWASAKI ENDOCARDITIS PERICARDITIS

CONNECTIVE TISSUE DISEASE: MARFANS LOEYES-DIETZ EHLER-DANLOS

CARDIOMYOPATHIES: HCM/HOCM DCM

COMMON GENETIC SYNDROMES ASSOCAITED WITH CHD
CHROMOSOMAL ABNORMALITIES 50-80% (1% IN FT)

DOWNS: (CAVC TOF DIGEORGE/VCF: (IAA TOF-PA/APV Truncus) test hypocalcemia

WILLIAMS: (SUAS PPS) remember ? vit D supplementation

ALAGILLE: (PPS)bil atresai

TURNER BAV/COA AAo anevr NOONANS: PS/HCM

CONGENITAL HEART DEFECTS…..

20 % VSD

5-10% ASD PS PDA TOF HPLHS

3-6% CoA AS TGA

2-3% PA-IVS Tri A

1-2% PA-VSD DORV Truncus TAPVC

0.5-1% Ebsteins CCTGA SV APwindow Cor Triatriatum

CLINICAL PEARLS

SHUNT PRECORDIUM +INFLOW RUMBLE indicates QP/QS >2:1 and need for surgery

Shunt vascularity on CXR = vx>bronchiole

BP gradient –cuff on RA LA and one leg and rotate attmt –note HR

Large VSD can result in FTT without loud murmur as PVR falls

Some ASD can enlarge with body growth

LARGE PDA - rapid CHF since cor perfusion occurs in diastole-Aodias P decreased-LVEDP increased and time in dias shortened

FEM PULSES-decreased indicates CoA INCREASED/bounding in CHF = PDA/AP window or AR

LOUDER(holosys) murmur =higher Pgrad ie small defect or outflow obstruction-(ejection murmur)

CLINICAL PEARLS …

Cyanosis=R-L shunt Recognition requires minimum of 3% desat Hb so delayed in anemia

TACHYPNEA with min cyanosis maybe DTGA -VSD or TAPVC

INFRADIAPHRAGMATIC TAPVC –obstructed gives small heart and "RDS" picture on CXR

VALVAR PS in infancy can progress from mild to severe or resolve in first 6 mo

BAV can develop first clinical sign in adulthood –endocarditis may be presenting symptom

40% of HPLHS have parent with BAV

HETEROTAXY =AMBIGUOUS SITUS WITH MIDLINE LIVER

COMPLEX MOSTLY CYANOTIC LESIONS RT ISOMERISM (bilat rt lungs) ASPLENIA +PS/PA

LT ISOMERISM bilat left sides)POLYSPLENIA and anomalous venous conn

Identify LA –receiving pulm veins narrow base hookshaped atrial appendage

RA –suprahepatic IVC + coronary sinus LIMBUS OF FOR OVALE triangular broadbased AA

LEFT MAINSTEM BRONCHUS TAKE OFF

INCIDENCE CHD high in- ISOLATED DEXTROCARDIA AV CONCORD and NRGV "pivotal "

ISOLATED DEXTROCARDIA WITH L-TGA

NORMAL D-LOOP morphological RV is RT HANDED and anterior

L-LOOP morphological LV is RT HANDED and anterior

COMPLICATIONS/LONGTERM FOLLOW UP POSTOP CHD

- TOF: PVR –chronic PR and Rvdilation VT AR (large ao root) small arm
- FONTAN : “single ventricle pathway” from HPLHS to Tri A to unbalanced AVC Gleen obstruction chronic hepatic congestion---cirrhosis PLE pulm AVM
- MUSTARD/SENNING: PV/SVC obstruction AFibFlutter
- ARTERIAL SWITCH : Su PS (LeCompte maneuver) Su AS and cor ostial narrowing

KAWASAKI

- >5DAYS FEVER
- CONJUNCTIVITIS nonpurulent
- ORAL erythema cracked lips strawberry tongue
- CERVICAL node
- RASH urticarial to scarlet fever
- EXTREMITIES swollen hands/feet and digits palmar erythema
- ESR CRP anemia urethritis arthritis
- HYDROPS gallbladder
- 8 DAYS window to treat IVGG 2 gm/kg/12 hrs ASA hi vs low dose

LONGTERM FOLLOW UP KAWASAKI

- Echocardiography at dx or hospital dc then at 6-8 weeks with CBC/CRP not ESR
- if CRP normal and echo neg for CAA at 6-8 weeks stop ASA and rtn for echo at 1 yr
- If small/ medium aneuvr or persistent cor dilation then follow on ASA yearly 2D no contact sports
- Large aneyr ASA plus Coumadin Q 3 -6 mo 2D

- ALL PATIENT COR COUNSELING RE BMI/BP /SMOKING
- Reasonable to start statin "early" if indicated

MAJOR CARDITIS CHOREA ARTHRITIS ERYTH MARG SUBCUT NODULES

MINOR fever arthralgia increased PR increased ESR /CRP asp test

Support evidence ASLO/ANTIDNAse titers

2 major or 1 major and 2 minor

antiAcarbohydrate peak 1mo post nl by 2 yrs stys pos in chro MR

Valve ds resolves in 70-80% if secondary prevention is maintained

ARF JONES 1944

LA benz PCN Q 28 DAYS

Sufadiazone 1 gm daily

CHOREA jack in the box milkmaids grip labile mood " ADHD" poor handwriting choreiform movements ST VITUS DANCE 3 MONTS POST INF

ENDOCARDITIS

- JANEWAY nontender eryth/hemorrh palms soles
- OSLER nodes tender red lumbs fingers/toes
- ROTH spots “fluffy” white eyeground spots
- +RF IN 50 %
- 70% in CHD/1/1280 hosp may occur in NN with normal hearts
- 80% STREP (viridans/alphahemo) or coag neg STAPH
- <10% GRAM NEG “HACEK “
- 5% NEG BLOODCX
- PCN GENT or CEF GENT if prosthetic valve VANCO GENT

PROPHYLAXIS GUIDELINES

HIGH RISK

1. PRIOR HX/OF ENDOCARDITIS …IVDA
2. PROSTHETIC VALVE /SHUNTS prosthetic material
3. UNREPAIRED CYANOTICS REPAIRED +post patch x6 mo +residual defect
4. TRANSPLANT with VALVE DS

MODERATE RISK AMOX /CLINDA/AZITHRO

1.BAV 2.MVP 3 RHD SLE 4 HCM

5.UNOP PDA VSD AVSD

INTRAUTERINE CIRCULATION AND CHANGES AT BIRTH

- CIRCULATION IN PARALELL –R-L ATRIAL AND DUCTAL SHUNT to fill left side and provide bypass of lungs
- Only 8 % combined cardiac output over isthmus and peripheral PA's – "coarctation watch" and physiological PPS murmur
- RISK FACTORS- TYPE 1 and 2 DIABETES PRIOR sib/parent with CHD
- Exposure lithium/anticonvulsants/antidepressants pesticides? Automechanic beautician
- At birth =separation from placenta/PGE + increase in arterial O2 leads to normal closure duct and FO----30-40% adults have PFO ? Chronic migraine – potential risk of paradox emboli
- Normal decrease in PVR over 6mo (delayed recognition large VSD /ALCAPA

DUCTAL DEPENDENT LESIONS

FOR SYSTEMIC OUTPUT	FOR PULMONARY BLOOD FLOW	MIXING
HPLHS	HPRHS PA Tri ATRESIA	DTGA
CRITICAL AS		
TIGHT COARCTATION	TOF-PA or severe PS	

ARRHYTHMIA

SVT

AVNRE

WPW ANTI/ORTHODROMIC

AFIB/AFLUTTER

NN AFLUTTER

ATRIAL ECTOPIC TACHYCARDIA

VT

WIDE COMPLEX –SLOWER RATE THAN SVT/WPW

CATECHOLAMINE SENS VT –bidir VT lethal in 30-50% bbloq ICD

SUSTAINED VT 50% NORMAL HEARTS

Repaired TOF HCM DCM ARVD

LQTS Electrolytes OD BRUGADA (RBBB and ST elevation)

IDIOPATHIC FASCICULAR LV T-reentry in LV =RBB +LAD or RBB +RAD

(DDX SVT with RBB resp to verapamil)

LQTS

ROMANO-WARD autosomal dominant

JERVELL-LANGE NIELSEN autosomal recessive associated with congenital deafness

Deafness/seizure d/o requires baseline ECG

CONGENITAL CAVB may present as arrhythmia –frequent Pac's

SUDDEN CARDIAC DEATH

- HCM 36%
- COR ARTERY ANOMALY –(LEFT from RT COR SINUS) 24 %

- MARFANS MVP AO ANEURYSMS 6 %
- CHD 5%
- DCM MYOCARDITIS ARVD 3%
- COR ARTERY DISEASE 2 %

- COMMOTIO CORDIS <1 %

HCM/HOCM

- INFANCY- IDM regress after 3mo NOONANS or GSD (infants + RVOTO)
- INCREASED MURMUR with sudden standing decrease with squatting
- Abnormal ECG may preceed symptoms
- 80 % abnormal intramural cor – myocardial bridge –inherent diastolic dysfxn
- Mortality rate 1% /yr adult 2%/yr in children
- Genetic marker known start screening early
- if fam hx of SCD follow echo until 18 then Q5yrs
- >30 mm IVS prob needs AICD
- Trained athlete with LVH-stop all exercise x 8 weeks and reecho

MATERNAL SLE

- SCREEN FOR CONDXN DS IN UTERO-

- Refer for fetal echo if mom has SLE/RA or known +SSA
- 18-28 WEEKS gestation follow mechanical PR interval

- Risk CAVB 1-8% if mom has SSA/SSB
- 15-18% if prior child with CAVB
- 60 % WILL REQUIRE PACEMAKER over lifetime

CARDIOVASCULAR RISK FACTORS

- OBESITY SEDENTARY LIFESTYLE
- PREDIABETES/INSULINRES ACANTHOSIS NIGRICANS
- NORMAL FAST TRIGLYCERIDES 40
- IDEAL TOTAL CHOL 150-160 LDL 90-100 HDL 50-55
- HYPERTENSION
- FAMILY HX/OF EARLY MI <55 Y/OLD
- KAWASAKI ARTERIAL SWITCH PA-IVS COR reimplantations TRANSPLANT
- SCREEN ALL >20 or earlier if risk factors
- TREAT if fails diet x1 yr + LDL>160 with fam hx + 10 y/old LDL >190 2 risk fct

- Congen RBBB and athletic heart ? Increased risk of AF

INDICATION FETAL ECHOCARDIOGRAPHY

- Prior child with CHD/Parent with CHD
- DM type 1 type 2
- Teratogenic exposure
- Known chromosomal fetal defect/ fetus with multiple anomalies
- Maternal SLE

ADCHD

INCREASING POPULATION more complex interventions –EP interventional cath

Need for management of coronary heart disease/other cardiovasc risk

Difficulties in employment increased risk of mental d/o

Need for management of pregnancy –fetal echocardiography MFM

ACHD IN THE UNITED STATES

- 2010 273,000 adult survivors projected to be at least 510,000 by 2050
  incidence of CHD of .9 % - mod to severe .6 % - 2% if BAV /AS/AR

. WORLDWIDE 3/million

Increasing number of survivors and more complex lesions to follow

impact of neurological sequelae after several OHS during first 2 years of life/employment opportunities

Complex arrhythmia-diff acces thru past surgeries-failing systemic ventricles..